VOL. XV. March, 1890. No. 3

THE
Journal
OF
Nervous and Mental Disease.

Original Articles.

NOTES ON SOME CASES OF CHOREA AND TREMOR.[1]

Case I.—Hereditary (Huntington's) Chorea in a Negro. Case II.—Hereditary (Huntington's) Chorea, Apparently with Absence of History of Heredity. Case III.—Hereditary Chorea with History of Chorea in Father and Infant Child. Case IV.—Paramyoclonic Chorea, with a History of Other Cases in the Same Family. Case V.—Tremor of Paralysis Agitans. Case VI.—Tremor of Disseminated Sclerosis. Case VII.—Tremor from the Abuse of Alcohol and Tobacco. Case VIII.—Diffuse Undulatory Tremor. Case IX.—Diffuse Undulatory Tremor.

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Reported by J. L. BOWER, M.D. Resident Physician.

IN the Philadelphia Hospital Wards for Nervous Diseases are many cases illustrating diverse forms of chorea and tremor. Some of these have already been reported; others are examples of types of disease which have been so thoroughly studied and recorded that details of them would probably not serve a useful purpose; although a complete investigation of all, with sphymographic tracings, and close descriptions of the peculiarities of movement, might constitute a valuable contribution.

[1] Presented to the Philadelphia Neurological Society, January 27, 1890.

Condensed notes will be given of a few cases with brief comments, particularly directed to the observed peculiarities of tremulous or choreiform movements. Of the nine cases here reported five have been observed in the Nervous Wards of the Philadelphia Hospital; the other four have been furnished me by Dr. Mills,from notes of cases observed elsewhere, One of the most interesting of the cases (Case IV.) has been in the Hospital for a considerable time and has been frequently studied and commented upon by the physicians in attendance. Dr. James Hendrie Lloyd gave a sketch of this case in a clinical lecture recorded in the *Medical and Surgical Reporter*, May 19, 1888; but it is sufficiently important to call for a more detailed report. Cases X. and XI. would seem to be practically undescribed forms of fibrillary tremor, as exactly similar cases are not to be found in the text-books. Dr. Mills has suggested to designate this form of tremulous movement *diffuse undulatory tremor*. The cases of paralysis agitans, disseminated sclerosis, and alcoholic tremor are introduced to compare the varieties of tremor.

CASE I.—Hereditary (Huntington's) Chorea in a Negro.

E. H., age 36, barber. His father died of some disease unknown to the patient. The disease from which he is suffering is inherited from his mother; she with two sisters and one brother were or are suffering from the same trouble. In his mother the disease developed at the age of 26 and gradually grew worse until she died at the age of 68 from exhaustion; he does not know exactly at what age it developed in his aunts and uncle, but thinks that it was somewhere between 20 and 35; one of the aunts is still living at the age of 50 years, the other is dead; the uncle is 42 years of age and in him the arms alone are affected.

The patient has been a hard drinker; when 28 years old he was drinking heavily and became suddenly paralyzed in both lower extremities and remained in this condition for three months, when power gradually returned. From description the affection does not seem to have been multiple neuritis, as the paralysis came on suddenly and he had no pain. Three years ago he had a venereal sore, probably a chancre, followed by a bubo.

Six years ago he first began to have twitching of the hands; this spread gradually but very slowly, until his

whole body became affected and his present condition reached. When he stands he sways a little from side to side, occasionally lifts one or the other foot, raises and depresses his shoulders, almost constantly opening and shutting and contorting his fingers, at times also thrusting one or both hands to his back. He is never entirely still, and he gives the impression that he is trying hard but unsuccessfully to control his nervousness. When he walks his gait is stiff, weak and unwieldy (perhaps in part from the old paralysis). The irregular movements in the hands and fingers continue, and in the extremities he has occasional swaying and pseudo-ataxic movements; now and then he lifts one foot high and balances himself before stepping out. When sitting down he is still unquiet, but in less degree; his trunk, arms and hands are moving almost continuously, the latter being shifted from his knees to his thighs where he places them in order to keep them quiet. All his movements consist of a series of irregular starts affecting one part after another or several parts together. At times they have the appearance of design, and the patient seems also to be trying to restrain them. The movements are not increased by voluntary efforts, but seem to be by excitement. He writes fairly well, but slowly and with difficulty, and it appears to be a great effort to keep himself in the proper position and direct his movements for writing. His signature is given below.

His speech is slow, and at times he pauses as if to coordinate the movements of articulation.

On inquiry it is found that the parents and ancestors of the patients were all full blooded negroes. The patient's mental condition so far as can be determined, is not impaired. His physique is good; height, five feet nine inches; weight, 130 pounds. His lungs, heart and kidneys are normal.

This case is interesting for several reasons. In the first place, it is another illustration of hereditary chorea. Its occurrence in a negro adds to the interest, as chorea of any type is rare in this race. His case presents points of great similarity to that of Miller, a patient in the same wards, an account of whose disease and remarkable family history

has been given by Dr. Wharton Sinkler, in one of the best papers on hereditary chorea.[1]

The next case, so far as the choreiform symptoms are concerned, would seem to be best classed as Huntington's chorea, but the most careful inquiries, could elicit no history of heredity. The patient's mental condition is somewhat impaired, but not to a marked extent, and he gives a clear history of the absence of nervous disease in his family. He would be able to recall the striking fact of a disease similar to his own having occurred in his family. His parents, brothers and sisters, were certainly not so affected.

CASE II.—Hereditary (Huntington's) Chorea, apparently with absence of history of heredity.

H. C., age 32, a painter, to the coming on of his present disease had excellent health. He is a moderate drinker, and denies venereal trouble ; he does not use tobacco. His family history is entirely negative.

About three years ago he began to have pain in the right knee which was swollen ; in about a month he recovered from this trouble, but shortly afterwards he noticed that his hands and his fingers were weak, and sometimes trembled or twitched ; his legs and head also became affected about three months after his upper extremities.

When sitting down his legs and feet are usually quiet, but he has a most constant though not very marked movement of his head, trunk, and upper extremities ; his head is moved a little backwards or from side to side in an almost rhythmical manner ; his trunk is occasionally lifted or twisted as if about to. shrug his shoulders ; he carries his left arm across his chest, the hand and fingers partially extended ; his right arm generally hangs by his side at full length ; both arms are kept comparatively quiet, but occasionally are shifted about uneasily ; his fingers and hands are constantly moving, going through a series of irregular or athetoid movements—perhaps one or two fingers are flexed while others are extended, or one or all fingers may be extended partially or fully ; sometimes the fingers are thrust apart or sometimes the thumb and fore-finger are brought together and then separated. If the hands are supported on the knees, the movements lessen considerably ; occasionally he shifts the position of his hands or moves his fingers up and down.

[1] Journal of Nervous and Mental Disease, February, 1889.

When he walks his choreiform movements are much increased. Athetoid movements of the fingers are especially marked. He walks with a sort of twisting or half rotating movement of the trunk and limbs, occasionally lifting one foot high in the air and bringing it down with a flourish, and every few steps pausing as if to poise himself. By a strong effort of the will the movements seem to be lessened. Voluntary effort such as writing, or conveying a glass of water to his lips, cause the movements to be less marked. Excitement of any kind greatly exaggerates his movements. He cannot protrude his tongue fully, and loss of power or control of the tongue seems to be of the same character as that shown in other parts of the body.

Knee jerk and muscle jerk are considerably exaggerated; slight ankle clonus is present on both sides. He has no loss of sensation. During sleep the movements cease entirely. His hand-writing is quite passable, but when he writes he seems to be putting forth extraordinary effort.

The following is his signature:

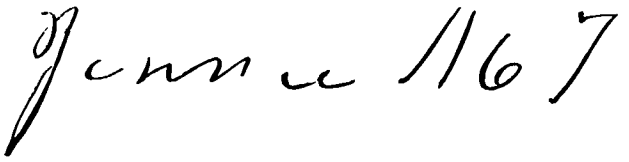
His speech is slow but quite intelligible; it seems to require an extreme effort to co-ordinate the movements of articulation. He can read fairly well by holding the book against his chest to steady it. His general health is excellent; height 5 feet 11 inches; weight 145 pounds; lungs, heart and kidneys are normal.

CASE III.—Hereditary chorea, with history of chorea in father and infant child.

The notes of this case are meagre, but it is worth while to report it as it adds another authentic record to the statistics which demonstrate the peculiar hereditary character of this form of chorea. The patient, J. M., married, white, was 41 years old at the time of coming under observation. Slight twitchings or choreiform movements began five years before, that is, when she was 36 years old. The disorder gradually increased. The movements were decidedly like those shown by the other cases of hereditary chorea reported in this communication. They affected the head, arms, legs, and speech, much more markedly on the right than on the left side. The patient ate well, her sleep was broken, and her mental condition was one of

slight beginning dementia. Her father was affected with the same or a similar disease. He was a hard drinking man. She had a baby only four months old, almost constantly affected with choreic movements of both hands.

Signature:



The following seem to be the peculiarities of movement and tremor in these cases of so-called hereditary chorea: The movements are usually first noticed in the upper extremities, especially in the hands. The disorder spreads slowly and gradually, but very surely, to other parts, until the entire body is more or less affected, the upper extremities commonly continuing to exhibit the symptoms in the most marked degree. Excitement increases, voluntary effort decreases the movements. When sitting or lying down the patient is much quieter than when standing or walking. Speech and writing areboth affected, and as the disease advances require great effort on the part of the patient. Only in its very early stages would this disease be likely to be confounded with any of the other forms of tremor or chorea. Very early it is conceivable that even disseminated sclerosis might be confounded with it; but early or late the the tremor of disseminated sclerosis is usually more confined to particular parts; and in disseminated sclerosis voluntary effort markedly increases the tremor, while it has the opposite effect in hereditary chorea. Emotion aggravates both disorders. When the head is affected in disseminated sclerosis the movement is a more or less uniform oscillation, while in hereditary chorea the head is moved about in an irregular manner—forwards, backwards, or to one side, as if the exertion was partly under the control of the patient's will.

CASE IV.—Paramyoclonic chorea, with a history of other cases in the same family.

C. E., aged 21, huckster, had three sisters who died of phthisis, at the ages of 4, 18 and 19 respectively. One brother died two years ago and was affected with the same disease as the patient, the age at which it developed and the causes not known. He has two brothers living, and he says that one of them has the same disease, only not so far advanced. In this brother the affection began at the age of 20 with no apparent cause, and has now existed for

over a year. His other brother is perfectly well. No history of nervous disease on either paternal or maternal side could be elicited. The patient says that before the advent of his present disease he was always healthy, never used tobacco, or intoxicating drinks, and had no venereal history.

Three years ago while huckstering his horse became frightened and ran away. When he found that the horse was beyond control he jumped from the wagon, hurt himself, and became unconscious; on coming to his senses his whole body jerked as it does at present although not so markedly. He was carried home and put to bed and remained there for five weeks, at the end of which time he was able to walk with the aid of canes, but frequently fell down. At present he is unable to walk and must be carried from the bed to his chair, in which he must be tied or he would be thrown to the floor by the excessive, inco-ordinate, involuntary, universal movements. He is unable to feed himself because of jerking of the arms. His appetite and general health are excellent; he sleeps well and during sleep is frequently quiet. It is impossible to obtain knee jerk as the limb becomes rigid when an attempt is made to elicit it. He has slight left ankle clonus. He has control o bowels and bladder. He shows no impairment of sensation or of the special senses.

The following is an attempt at a description of the character of the choreiform movements: When lying quiet, not watched or spoken to, or excited in any way, he is comparatively easy and may be perfectly quiet for several minutes or even much longer. Occasionally he will raise one or both hands slightly or it may be place them on his breast, or he may flex and extend the fingers in an irregular manner. These movements are made slowly and at intervals, but usually from six to twelve in a minute. Excitement or emotion, or attempted voluntary effort, greatly increase the movements and they rapidly become general; they are violent, irregular, inco-ordinate, wild, and shock-like; they are more or less independent of each other, numbering from sixty to eighty per minute. During the excitation the body is thrown forward and backward, the head is moved from side to side and often thrown forward and backward quite strongly. The facial muscles are usually free except theorbicular muscles of the eyelids. His upper extremities move rapidly, and usually irregularly and independently of each other. With the irregular movements he has at times certain automatically repeated acts. The most frequent of these seems to be a brushing of the head and face with the hands either open or closed, or a slapping

of one hand in the other, or on the knee. Frequently he throws his arms out at full length, then rapidly across his chest and face; or he may clasp his hands for a moment or execute some other irregular movement in the lower extremities, not so marked as in the upper, but otherwise very similar. He flexes and extends his thighs, legs, and feet in the most irregular manner; occasionally his legs are extended perfectly straight for a few seconds in what appears to be a tonic contraction, but it is due to a very rapid clonus. When he is lifted from his bed his body becomes rigid as a board; his arms are thrown wildly about, and his lower extremities become almost immobile by the extreme rapidity of the muscular contractions of the body and lower extremities. When he is placed on his chair the violent contractions jump him up and down for a minute or two. His speech is interfered with apparently through difficulty in co-ordinating the muscles of articulation, but when he commences to speak he talks quite rapidly.

The mental condition of the patient seems to be unaffected by the disease, and his general physical condition is good.

A careful consideration of this case shows points of great similarity between it and cases of hereditary chorea, although at first sight this resemblance might not attract attention. The movements are similar although far more violent and general. The history shows other cases in the same family. The case differs from hereditary chorea in having begun acutely, and at an earlier age than is common in the latter disease; also in the much greater rapidity and violence of the inco-ordinate movements.

CASE V.—Tremor of Paralysis Agitans.

A. K., age 54, a weaver, for sixteen years has been troubled more or less with chronic rheumatism, principally in the shoulder-joints. He is a very moderate drinker and smoker, and has no history of venereal troubles.

About five years ago he first noticed a weakness in the right arm, and this has steadily progressed. Three years ago a tremor of the right hand and fingers first became manifest; at first it was not constant and was more marked when under excitement. About one year ago the right foot "trembled" also, and within a month the left hand developed a fine tremor which is not always present. This tremor is fine and rhythmical, and never ceases except during sleep. By an effort of the will it can be lessened but not stopped; voluntary effort as in drinking from a glass, very much lessens it; excitement increases it con-

siderably. Grasping the arm lessens it decidedly but does not cause it to cease. If the hand is rested on the knees the tremor continues but is less marked. There is slight stiffening of the right hand, and he writes slowly and with considerable effort, but comparatively well.

The patient's shoulders are stooped, the head carried toward the chest and held fixed as if in a stock. The face is fixed, dull, and expressionless. He shows nothing peculiar in his gait. Sensation is not impaired.

Signature :

Several cases of paralysis agitans, more typical, or at least more advanced, than the one here reported are in the Nervous Wards of the hospital, but this case was selected for report and comparison because at a stage, so far as the tremor is concerned, when, if ever, it might be confused with some other affection.

In the main, this case affords the diagnostic features usually given by the books; the tremor is lessened by an effort of the will; it is, however, increased by excitement. It is fine and rhythmical, and fixity of limb and feature are present.

CASE VI.—Tremor of Disseminated Sclerosis.

J. O., age 71, laborer, ten years ago had mild malarial and four years later break bone fever, both of these attacks persisting for about five months. Up to ten years ago he was a hard drinker. He is a heavy user of tobacco, both chewing and smoking. He has no history of venereal troubles.

Two years ago he noticed a tremor of the right hand which first became apparent when he attempted to use the hand as in eating; this gradually became so marked that when possible he rested his right hand and used the left one. In about two months he noticed that the left hand also trembled, and shortly after he felt the same tremor in his body and lower extremities, although not so marked.

When the patient sits with his hands on his knees they are comparatively steady, but occasionally a fine tremor is seen. When his hands are extended without support, the

tremor is quite marked in both but more apparent in the right hand. When he attempts to control his movements by mental effort, the tremor is considerably exaggerated. If the hands or arms are grasped the tremor continues, and can be felt involving the entire linb. A marked increase occurs if a voluntary effort is made, as when he attempts to drink from a glass—the tremor then becomes wide, coarse, irregular and jerky, and in efforts to carry the glass to his mouth much water is spilled. There is a fine oscilation of the head, chiefly in a lateral direction, and occasionally a slight spasm or twitching of the zygmatic muscles of the right side of the face. The tremor involves the trunk slightly; he has also tremor of the tongue.

Nothing particular is noticed about his gait except that it resembles that of paralysis agitans in that his head is slightly bowed; his face has rather a blank and listless expression, but this may be accounted for by his age and low intelligence; he complains of weakness of both upper and lower extremities, especially of the right arm. The dynamometer shows a grasp of 26 for the right, and of 56 for the left hand. The least exertion fatigues him. This weakness first became manifest seven months ago and is steadily growing worse. There appears to be no impairment of sensation; the pupils are equal, moderately dilated, and respond to light; he has no nystagmus. Knee jerk is abolished and slight ankle clonus is present on both sides. He is frequently troubled with headache and vertigo, the latter being most marked in the morning. He speaks in a slow and tremulous manner. He has perfect control of the bladder and rectum.

CASE VII.—Tremor from abuse of tobacco and alcohol.

J. R., aged 63, about a year ago began to be afflicted with tremor in both hands, which has become gradually worse during the last three months. He complains of feeling dizzy, and if he turns suddenly is in danger of falling. He has no headache, nor any pain in the body. He sleeps well but has no appetite. For the past thirty years he has been accustomed to drink several glasses of beer and smoke twelve pipes during a day.

The tremor affects both hands about equally, it is coarse but not large, and keeps up continually without any variation; it is sometimes increased by exertion. The arms as well as the hands are somewhat affected. Sometimes the tremor almost entirely disappears for one or two days; and in the latter part of the day is generally better than in the morning.

Under enforced abstinence from alcoholic drinks and

tobacco, in about two months this man's tremor almost absolutely disappeared. The peculiarities of the tremor were its bilaterality, its complete discontinuance at intervals, its greater regularity than in disseminated sclerosis, its better response to treatment, and the absence in the patient of the other symptoms and conditions which are usually associated with paralysis agitans or disseminated sclerosis.

CASE VIII.—Diffuse undulatory tremor in a case of chronic spinal degeneration.

C. J. H., age 53, had had some pain in the lower part of the back for years, after which he noticed trouble with his feet when walking down hill. He grew gradually worse, but had no history of true pain, only cramp or spasm in the calves.

He cannot lift his toes in walking, stumbles and feels his feet under him. He has no trouble with his bladder or bowels. Twelve years ago he had some confusion of sight, but was relieved by glasses; he has no diplopia. The knee jerks are exaggerated. He has advanced paresis of both legs. The back muscles are not rigid. In the arms he has no palsy but a jerking or twitching of the muscles. For four months he has had cramps in the legs, and in the arms, but not so marked; also a dragging sensation on the left side of the lower part of the abdomen. He has, at times, a tingling sensation all over his legs, and sometimes in his arms. He has "sore" feelings in the thighs and legs.

A marked and very peculiar feature in this case is an almost universal tremor of the muscles of the thighs, legs, arms, forearms and trunk. Testing him with electricity it was thought at first that the tremors were contractions produced by the electrical application. Wave-like movements are present all the time. The surface of the body presents the appearance of a gently undulating sheet of water. In the shoulders, arms, etc., almost every physiological movement of the muscular groups could be seen; the muscles could be picked out by these involuntary contractions.

CASE IX.—Diffuse undulatory tremor in a case of chronic spinal degeneration.

G. B., 31 years old, white, married, two years before coming under observation noticed that his left leg began to fail, and six months later his right became similarly affected. In a few months the loss of power in the left lower extremity was very extensive; at the time of examination all the foot movements being abolished and those of the leg and thigh much weakened. The right leg and the left upper extrem-

ity paretic, but still retain considerable power. Knee jerk on each side are retained, muscle jerk increased; farado-contractility is retained, but somewhat diminished in some of the atrophied muscles of the left leg. Sexual power is good; he has no affection of the bladder or bowels and no anæsthesia.

The marked peculiarity of this as of the last case is the presence of a diffuse undulatory tremor, affecting both legs very generally.

The tremor in these cases is similar to that observed in the tongue in some bulbar strophic cases, and doubtless its pathology is the same as in these, but it is very rare to observe this extreme and widespread undulatory muscular tremor affecting parts other than the tongue.

EXOPHTHALMIC GOITRE.

Before a branch of the British Medical Association, reported in the *British Medical Journal*, Jan. 4, 1890, Dr. Cheadle showed a case of exophthalmic goitre in a male subject, in whom recovery was maintained for more than twenty years. He considered the symptoms due to disorder of the medulla, of the upper cervical portion of the cord, the region of the cardio-inhibitory, accelerator, vasomotor, vomiting, and glycogenic centres, all of which were—some constantly, some occasionally—involved in the disease. In February, 1868, the patient suffered from characteristic vascular excitement, marked exophthalmia, enlarged thyroid, which pressed so severely on the trachea that the consequent dyspnœa compelled the patient to sleep sitting in a chair. Tincture of iodine was freely given, followed by improvement in the course of a week, and this had continued and steady. The tendency of the disease seemed to be toward slow recovery in from one to five years. Cases were on record from death from pressure on the trachea, from acute mania, from cerebral hæmorrhage, from pulmonary congestion and anasarca, and from various intercurrent affections. One of the greatest dangers Dr. Cheadle considered to be persistent vomiting with diarrhœa. Absolute rest was essential in the acute stage, and opium with digitalis or belladonna. Galvanism was useful in the chronic state, but caused undue excitement in the acute form. L. F. B.